Sarcoidosis

DESCRIPTION

Sarcoidosis is a diverse systemic disease, characterised by the development of epithelioid granulomas and other changes in multiple organ systems, with the lungs most often affected. Its etiology is considered idiopathic, although it may be a response to an unidentified organism or antigen. Clinical disease occurs when granulomata affect the normal architecture and function of involved tissues.

SYSTEMIC SIGNS

- Lung involvement (about 90 per cent): Chest X-ray (CXR) may show bilateral and symmetrical hilar lymphadenopathy and pulmonary infiltration. Symptoms may include difficulty in breathing (dyspnea), fatigue and cough
- Skin lesions (about 30 per cent), including erythema nodosum (red nodules on legs or arms) and cutaneous granulomata
- Ocular signs (about 20 per cent): see below
- Nervous system (about 10 per cent)
- Other, for example, liver, spleen, lymph nodes, salivary glands, joints, heart, muscle and bones.

OCULAR SYMPTOMS

These include a red eye, blurred vision, pain, photophobia and lacrimation.

OCULAR SIGNS

Uveitis

- Most often a unilateral anterior granulomatous uveitis, with mutton fat keratic precipitates. However, the uveitis may be bilateral or non-granulomatous
- Vitritis, with fluffy white infiltrate opacities ('string of pearls'), may be present
- Busacca nodules on the iris surface
- Koeppe nodules on the pupillary margin.

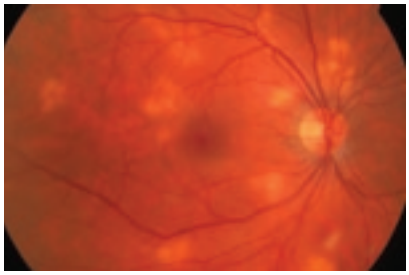
Glaucoma secondary to uveitis

Anterior ocular signs may include

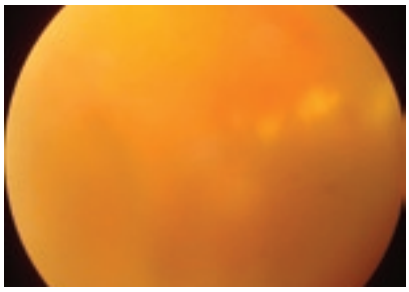
- Lacrimal – gland disease (keratoconjunctivitis sicca) in 20-30 per cent of patients
- Conjunctival slightly raised yellow lesions
- Band keratopathy.

Posterior signs are usually bilateral and may include

- Periphlebitis: white exudations affecting the equatorial retinal veins



Sarcoidosis showing pale lesions due to granulomas in the choroid and outer retina



Active vitritis with obscuration of retinal details

- Retinal vein occlusions, where granulomas may compress the vasculature
- Retinal haemorrhages
- Chorioretinitis
- Macular oedema – cystoid
- Optic disc oedema or granulomas
- Neovascularisation affecting the choroid, optic disc or peripheral retina.

Neurological effects

Visual pathways may be affected by granulomas, causing visual field restriction, facial nerve palsy, papilloedema.

The orbit or extraocular muscles

These muscles may also be affected by granulomas, causing diplopia, palsies or proptosis.

PREVALENCE

Occurs most frequently in adults (20-40 years), particularly women and darker skinned races. Uncommon to rare.

SIGNIFICANCE

Sarcoidosis is potentially vision and life-threatening if uncontrolled.

DIFFERENTIAL DIAGNOSIS

Lymphoma – primary intraocular (large reticulum cell sarcoma).

SEE ALSO

Syphilis, HIV, Lupus, Lyme, Sickle cell disease.

MANAGEMENT

Additional investigations

Diagnosis of sarcoidosis is usually assisted by chest X-ray. Angiotension converting enzyme (ACE) is usually elevated. Also consider biopsy of nodules in the lacrimal gland, conjunctiva, skin, lymph node or lung. Fluorescein angiography may be indicated to confirm posterior segment neovascularisation.

Laboratory tests

Blood tests are indicated for autoimmune, rheumatologic, infectious and inflammatory diseases, especially in the presence of atypical uveitis or other anomaly.

Topical and oral medications

Uveitis and sarcoid retinopathy are treated with topical or oral steroids; occasionally additional immunosuppressants are needed. Initial treatment may be prednisolone acetate 1 per cent q1h to q6h. Consider tear supplements and punctal occlusion if dry eye is present.

Laser treatment and injection

Photocoagulation may be indicated for posterior segment neovascularisation. Choroidal neovascularisation may be treated with repeated doses of anti-vascular endothelial growth factor (anti-VEGF) drugs delivered by an intravitreal injection.

Review

Routine review as part of a multidisciplinary approach is essential. Asymptomatic patients need six-month reviews for uveitis, cataract, glaucoma, dry eye or neurological signs.

Prognosis

Although sarcoidosis may remain stable for some patients, in others it tends to follow a waxing and waning course. Good vision can be expected if ocular inflammation or retinopathy can be kept under control.

The full series of these articles is available in the book *Posterior Eye Disease and Glaucoma A-Z* by Bruce AS, O'Day J, McKay D and Swann P. £39.99. For further information click on the Bookstore at **opticianonline.net**

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